CME

Hypertrophic Cardiomyopathy: A Review

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Hypertrophic cardiomyopathy (HCM) is a relatively common disorder that anesthesiologists encounter among patients in the perioperative period. Fifty years ago, HCM was thought to be an obscure disease. Today, however, our understanding and ability to diagnose patients with HCM have improved dramatically. Patients with HCM have genotypic and phenotypic variability. Indeed, a subgroup of these patients exhibits the HCM genotype but not the phenotype (left ventricular hypertrophy). There are a number of treatment modalities for these patients, including pharmacotherapy to control symptoms, implantable cardiac defibrillators to manage malignant arrhythmias, and surgical myectomy and septal ablation to decrease the left ventricular outflow obstruction. Accurate diagnosis is vital for the perioperative management of these patients. Diagnosis is most often made using echocardiographic assessment of left ventricular hypertrophy, left ventricular outflow tract gradients, systolic and diastolic function, and mitral valve anatomy and function. Cardiac magnetic resonance imaging also has a diagnostic role by determining the extent and location of left ventricular hypertrophy and the anatomic abnormalities of the mitral valve and papillary muscles. In this review on hypertrophic cardiomyopathy for the noncardiac anesthesiologist, we discuss the clinical presentation and genetic mutations associated with HCM, the critical role of echocardiography in the diagnosis and the assessment of surgical interventions, and the perioperative management of patients with HCM undergoing noncardiac surgery and management of the parturient with HCM. (Anesth Analg 2015;120:554–69)

Hypertrophic cardiomyopathy (HCM) was first described in 1868,[1] its functional consequences in 1957,[2] left ventricular (LV) asymmetric and especially septal hypertrophy in 1958,[3] and its familial nature in 1960.[4] HCM is a relatively common inherited disorder, with a prevalence of 1:500,[5] which is equivalent to at least 600,000 people affected in the United States.[6] HCM is also associated with Pompe, Danon, and Fabry disease and LV hypertrophy (LVH) due to mutations in AMP-activated protein kinase, mitochondrial abnormalities, and genetic dysmorphic syndromes, for example, Noonans, Kearns-Sayre, and LEOPARD syndromes.[5]

Surgical resection was first described by Morrow and Brockenbrough in 1961 but first performed by Cleland in 1958.[7–9] HCM has several synonyms, including hypertrophic obstructive cardiomyopathy, idiopathic hypertrophic subaortic stenosis, muscular subaortic stenosis, and asymmetric septal hypertrophy.[10] The goals of this review are to (1) outline the pathophysiology, clinical presentation and consequences, assessment, and medical management in patients with HCM; (2) discuss the role of echocardiography in HCM; (3) discuss treatments, important intraoperative echocardiographic indices, and the influence of surgery on long-term outcomes; (4) outline the principles underlying the management of patients with HCM who are undergoing noncardiac procedures; and (5) review HCM as it pertains to the parturient.

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PATHOPHYSIOLOGY AND CLINICAL PRESENTATION

HCM is defined as abnormal LV thickening without chamber dilation that is usually asymmetrical, develops in the absence of an identifiable cause (e.g., aortic valvular stenosis [AS], hypertension), and is associated with myocardial fiber disarray.[11,12] HCM is inherited as an autosomal dominant trait with variable expression. Defects of at least 11 genes and >1440 mutations demonstrate its genomic heterogeneity.[11,13,14] Mutations involving myosin heavy chain (*MYH7*) and myosin-binding protein C3 are most common, together accounting for 75% to 80% of sarcomere mutations in HCM. Genotype analysis can identify family members with the mutation (genotype +), even if they have not yet developed clinical signs (LVH–).[15]

The major underlying structural abnormalities in HCM are (1) myocardial cell disarray where the cells are in a disorganized pattern as opposed to a normal parallel myocyte arrangement; (2) coronary microvasculature dysfunction by increased wall/lumen ratio; and (3) remodeling changes.[16,17] In intramyocardial arterioles <80 μm, studies have revealed a 2-fold increase in wall-to-lumen ratio, predisposing patients to silent myocardial ischemia, ongoing myocardial injury, and fibrosis.[17] Moreover, these changes are not limited to areas of LVH and myocardial remodeling that occur as a compensatory mechanism and can involve changes to the myocytes, fibroblasts, and interstitium. These changes evolve for years before the onset of symptoms.

Disorganized myocyte pattern, increased wall/lumen ratio of coronaries, and remodeling changes in HCM patients lead to impaired coronary reserve, diastolic dysfunction, supraventricular and ventricular dysrhythmias, and sudden death. LV remodeling can include fibrosis, diffuse, asymmetric, focal or concentric hypertrophy, as well as decreased cavity size.[18]

LV outflow tract (LVOT) obstruction (LVOTO) occurs in HCM and it was initially thought that the basal septal

hypertrophy encroaching on the LVOT caused the obstruction. However, more recent studies have shown that during ventricular systole, flow against an abnormally positioned mitral valve (MV) apparatus results in drag forces on part of the leaflets, which are then pushed into the LVOT.[6,11,19–21] The MV apparatus abnormalities can include anterior displacement of papillary muscles, hypertrophied papillary muscles touching the septum, elongated mitral leaflets, or anomalous insertion of the papillary muscle onto the anterior mitral leaflet.[18,21,22] LVOTO can be precipitated or aggravated by decreased end-diastolic volume or systemic arterial resistance or increased contractility or heart rate.[23]

Patients with HCM can present early in life with symptoms or can live for decades asymptomatically. The most frequent symptoms include exercise intolerance, angina, dyspnea, dizziness, syncope, and/or sudden death.[18,24] In a cohort study by Spirito et al.,[25] the incidence of unexplained or neurally mediated syncope was 15% and was shown to be associated with a 5-fold increased risk of sudden death. Physical examinations may be normal at rest but may reveal a systolic murmur in the presence of functional LVOTO. Electrocardiography (ECG) changes include left atrial (LA) enlargement, ST segment depression, pathologic Q waves, and inverted T waves in at least 2 or more consecutive leads (Table 1).[26] The 12-lead ECG is abnormal in 75% to 95% of HCM patients.[6] Clinically, it may be difficult to distinguish an HCM patient from those with fixed AS. Although HCM can cause a dynamic LVOTO and AS is a fixed lesion, both HCM and AS patients can present with the aforementioned symptoms. An echocardiogram will be useful in these cases and provide information regarding severity and classification of the HCM and differentiate it from fixed AS (Fig. 1, A and B).

HCM patients can be classified in 3 categories based on their degree of LVOTO: "nonobstructive," "labile," or "obstructive at rest" (Table 2). About one-third of patients will have obstruction at rest (peak gradient >30 mm Hg), and one-third will have labile obstruction (peak gradient >30 mm Hg only during provocation).[6,10,27,28] Maneuvers used to provoke LVOTO for diagnostic purposes include the Valsalva maneuver, administration of a potent inhaled vasodilator, such as amyl nitrite, and exercise treadmill testing.[6] Another one-third will have no obstruction under provocation or resting conditions (peak gradient <30 mm Hg). Some nonobstructors demonstrate diastolic dysfunction and thus may have symptoms of exertional dyspnea.[29] Most patients with nonobstructive HCM do not develop severe progressive heart failure during their clinical course.[13] A new subset of genotype-positive, phenotype-negative family members has emerged in which the clinical ramifications are yet unknown.[14] This new subset is asymptomatic and has normal LV wall thickness but has a genetic mutation associated with HCM.

Table 1. Nonspecific Electrocardiogram Changes Associated with Hypertrophic Cardiomyopathy

-Left ventricular hypertrophy (S wave in V1 ≥35 mm; R wave in V5 >35 mm)
Left axis deviation/left anterior hemiblock
Intraventricular conduction delay (QRS >0.12 ms)
-Left atrial enlargement (broad notched P wave in lead II; deeply inverted P wave in V1)
-Pathologic Q waves
-Poor R wave progression in precordial leads
-Supraventricular arrhythmias (most commonly atrial fibrillation)
-Complete bundle branch block
-ST segment depression
-Inverted T waves in ≥2 consecutive leads

ECHOCARDIOGRAPHY AND OTHER IMAGING IN HCM

Echocardiography was first used to aid diagnosis in HCM in 1969.[30] Forty years later, echocardiography continues to be critical in the evaluation of patients with HCM. The American Society of Echocardiography recently described how 10 echocardiographic criteria/indices could be used in the diagnosis and treatment of HCM[31] (Table 3). Emerging echocardiographic techniques (e.g., tissue Doppler [TDI] and deformation analyses), as well as magnetic resonance imaging (MRI) and computed tomography imaging, are being used increasingly to evaluate patients with HCM. The latter (MRI and computed tomography) provide excellent anatomic information but are less useful in defining hemodynamic perturbations. TDI measures the velocity of myocardial movement in systole and diastole and have demonstrated that often there is a reduction in systolic and diastolic velocities even before the development of LVH.[31] The essential elements of an echocardiographic evaluation in HCM patients include assessments of ventricular dimensions, LVOTO, systolic and diastolic function, and MV function and geometry. Most initial evaluations involve transthoracic echocardiographic (TTE) imaging with the patient resting, during treadmill exercising, and during administration of amyl nitrite.

LV Wall Thickness

Although any myocardial segments can be affected, the septum is involved in approximately 90% of cases.[19] The prototypical findings are unexplained septal wall thickness of >13 mm (>15 mm in hypertensive patients[19] with a ventricular septal to posterior wall thickness ratio >1.3 in the LV).[32,33] Wall thickness can be best measured by 2-dimensional (D) TTE in the parasternal short-axis, apical 4-chamber, or parasternal long-axis view at end-diastole or by 2D transesophageal echocardiogram (TEE) in the transgastric short-axis view.[31,34] These measurements have prognostic significance. Spirito et al.[35] found that the risk of sudden death was 0 per 1000 person-years for patients with a wall thickness <15 mm (95% confidence interval 0–14.4) but increased to 18.2 per 1000 person-years for patients with a wall thickness of >30 mm (95% confidence interval 7.3–37.6).

Turer et al.[34] categorized subtypes of different phenotypes of HCM patients (Table 4) based on where their septal wall thickness was located and found a striking association between one particular subtype and certain clinical characteristics. Patients with the catenoid morphology, predominant thickening of the mid-septum, were found to have the youngest age at HCM diagnosis, larger LV mass and a higher prevalence, and extent of late gadolinium enhancement by cardiac magnetic resonance (CMR).[34] Gadolinium is a contrast medium used in MRI to enhance imaging, and specifically, late gadolinium enhancement in CMR detects myocardial fibrosis. This finding has prognostic value in predicting adverse cardiovascular events among HCM

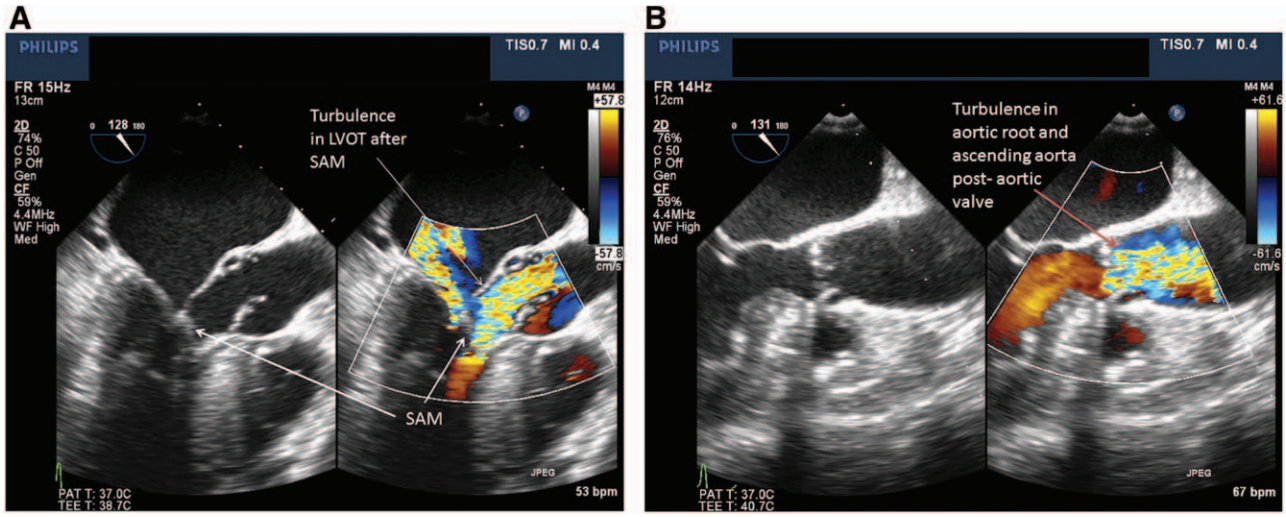

Figure 1. A, Transesophageal echocardiography (TEE) image—Color-flow Doppler (CFD) of mid-esophageal long-axis (ME-LAX) view in hypertrophic cardiomyopathy patient with turbulence in left ventricle outflow tract (LVOT) at the level of systolic anterior motion of the mitral valve (SAM) (proximal to the aortic valve). B, TEE image—CFD of ME-LAX view in a patient with valvular aortic stenosis demonstrating laminar flow in LVOT and turbulence distal to the diseased aortic valve.

patients.[36] Olivotto et al.[37] reported an association between LV mass determined by CMR and higher rates of HCM-related death. Patients with catenoid morphology had a higher percentage of New York Heart Association (NYHA) class 3 or 4 symptoms and restrictive (grade 3) diastolic dysfunction.[34]

Dynamic LVOTO

LVOTO in HCM is multifactorial and dynamic. In a real-time 3D echocardiography prospective study using 3D MV analysis on HCM patients with LVOTO, multiple linear regression analysis for independent determinants of minimal LVOT systolic area included end-systolic LV volume, total mitral leaflet area, interpapillary muscle distance, annular height, and LVOT hypertrophy index defined as the percentage area measuring >16 mm thick in the LVOT region[38] and was inversely related to LVOT pressure gradients (R^2 = 0.83, P < 0.001).[38] Turer et al.[34] found that in their classification of different septal morphologies, the catenoid and neutral septal subtypes had the highest resting gradients.

Under normal anatomic and physiologic conditions, the LV cavity has both an inflow and an outflow compartment. Inflow from the LA across the MV during diastole occurs predominantly posteriorly in the LV, while outflow during systole occurs anteriorly after the MV closure. The separation of these functional compartments in the LV and the normal closure and coaptation of the MV leaflets posteriorly away from the LVOT are determined by several anatomically interrelated variables. Importantly, these include ventricular cavity size relative to the size and location of components of the mitral apparatus.

A small ventricular cavity, a coaptation (C)-sept distance <2.5 cm, an anterior to posterior MV leaflet height ratio of <1.3, posterior annular calcification, mid-septal hypertrophy, and anteriorly positioned papillary muscle are all risk factors for LVOTO (Fig. 2).[19,21,34,38,39]

Table 2. Definition of Dynamic Left Ventricular Outflow Tract Obstruction

Hemodynamic state	Conditions	Outflow gradients[a]
No obstruction	Rest	<30 mm Hg
	Physiologically provoked	<30 mm Hg
Labile obstruction	Rest	<30 mm Hg
	Physiologically provoked	≥30 mm Hg
Obstruction at rest	Rest	≥30 mm Hg

[a]Gradients are the peak instantaneous continuous wave Doppler gradient.[6]

Table 3. Echocardiographic Focus in Hypertrophic Cardiomyopathy

1. Presence of hypertrophy and its distribution; report measurements of LV dimensions, wall thickness (septal, posterior, maximum)
2. LV ejection fraction
3. RV hypertrophy and presence of RV dynamic obstruction
4. LV volume indexed to body surface area
5. LV diastolic function (relaxation and filling pressures)
6. Pulmonary artery systolic pressure
7. Dynamic obstruction at rest and with Valsalva, site of obstruction and gradient
8. Mitral valve and apparatus evaluation, details of mitral regurgitation (i.e., mechanism, severity); TEE if needed
9. TEE to guide surgical myectomy and TTE/TEE for alcohol septal ablation
10. Screening

LV = left ventricular; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.[31]

Doppler echocardiography is critical in determining the degree of LVOTO by measuring the increased velocity of blood flow at the site of obstruction. Spectral Doppler is used to determine both the anatomic site of obstruction (pulse wave Doppler) and the peak gradient across the LVOT.[10] Color-flow Doppler can also assist in distinguishing the site of obstruction due to the increased turbulence seen distal to the obstruction.

In HCM patients with hemodynamically significant lesions, Doppler interrogation of the LVOT reveals a

Table 4. Septal Morphology in Hypertrophic Cardiomyopathy Patients and Transthoracic Echocardiogram Views

Simple sigmoid	Maximal septal wall thickness located in basal septum by both LAX and apical 4-chamber (A4) views
Catenoid	Maximal septal wall thickness in mid-septum by A4 views
Neutral	Uniformly thickened septum (basal, mid-, and apical) by A4 view
Apical	Maximal septal wall thickness in apical septum by A4 view

LAX = long axis.[34]

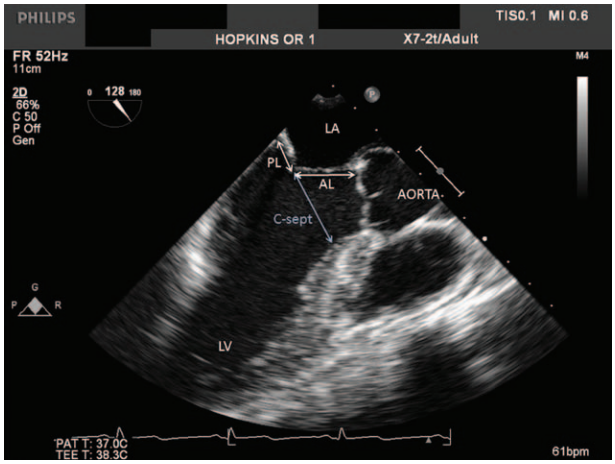

Figure 2. Transesophageal echocardiography mid-esophageal long-axis view. AL = anterior leaflet; PL = posterior leaflet; AL length/PL length (as indicated by the arrows) = anterior to posterior MV leaflet height ratio; C-sept = coaptation-septal distance; LA = left atrium; LV = left ventricle.

late-peaking dagger-shaped velocity tracing consistent with dynamic and increasing LVOTO occurring in mid- to late systole[40,41] (Fig. 3A). This is due to progressive encroachment of the anterior MV leaflet into the LVOT that causes a progressive decrease in the cross-orifice locally during systole and contrasts with a fixed obstruction secondary, for example, to aortic valve stenosis (Fig. 3B).

Systolic Function

Systolic function, as determined by ejection fraction (EF) or fractional shortening, is usually normal or slightly increased in both obstructive and nonobstructive HCM. Importantly, significant hypertrophy can lead to small LV end-diastolic volumes, which can reduce stroke volumes, despite a normal EF.[11,31] In a subgroup of HCM patients (2%–5%), right ventricular and LV systolic function may deteriorate as the disease progresses, a result of myocardial fibrosis.[42] Myocardial wall thinning, increased end-systolic volume, and moderate ventricular dilation may ensue. A resting EF <50% (end-stage HCM) is associated with an increased mortality (11% per year), including risk of sudden death.[42]

TDI and strain are newer techniques that can be used to evaluate systolic function. TDI measures the velocity of myocardial tissue using low-pass filters to screen out higher velocities created by blood flow. Strain measures segmental myocardial deformation, and strain rate measures the rate of this deformation. Using these techniques, significant contractile impairment may be detected despite an overall preserved EF.[43] Studies have demonstrated decreases in strain patterns in HCM patients compared with controls.[44,45] Moreover, strain rate imaging has been used to distinguish LVH secondary to hypertension from that occurring in nonobstructive HCM.[43,46] Kato et al.[43] found that a combination of interventricular septal wall thickness to posterior wall thickness ≥1.3 and systolic strain of ≤−10.6% was able to discriminate HCM from LVH due to hypertension with a predictive accuracy of 96.1%. Although strain is supposedly a systolic factor, the tissues' recoil ability, controlled by LV end-systolic volume, is important in the rate of relaxation. Therefore, systolic strain reflects the lusitropic ability of the myocardium and, thus, LV end-diastolic pressure. Importantly, reduction in strain and strain rates is seen in HCM patients before LVH, ECG abnormalities, or focal areas of fibrosis are detected.[47]

Diastolic Function and LA Evaluation

Impaired relaxation is common in HCM patients. Interstitial fibrosis and increased LV mass contribute to increased myocardial stiffness and decreased chamber compliance. Moreover, HCM patients without obstruction may actually have more severe diastolic dysfunction compared with HCM patients with obstruction.[29] Diastolic dysfunction can result in LA enlargement and is associated with exercise intolerance and poor outcomes in HCM, mainly due to supraventricular arrhythmias.[48–52] Markedly increased (>48 mm) anterior-posterior LA diameter in the mid-esophageal 2-chamber view on 2D-TEE is seen with chronic mitral regurgitation (MR) but can also be indicative of diastolic failure[53] and is associated with increased risk of death in HCM patients.

MV Findings

LVOTO in HCM occurs due to systolic anterior motion (SAM) predominantly by the hydrodynamic force of drag and the pushing force of flow (Fig. 4). Flow-drag-flow forces occur when anterior displacement of the papillary muscles cause diastolic inflow to be directed toward the septum (as opposed to posteriorly), and thus, outflow is directed posteriorly, causing drag forces on the mitral leaflets into the LVOT as blood is ejected (Fig. 5). Historically, it was felt that the venturi mechanism, whereby high velocity in the outflow tract lifts the MV toward the septum, was the main cause of SAM.[54] This theory was disproven by several studies that demonstrated that SAM begins early in systole at a time when the LVOT velocity is normal.[21,55–58] Flow drag is the component of force on a body that is in the direction of flow, as opposed to "lift," which is venturi forces produced by high-velocity flow over the surface of an object causing the object to move perpendicular to flow. Decreased velocity significantly decreases lift in 2 ways and attenuates its impact compared with drag. Lift

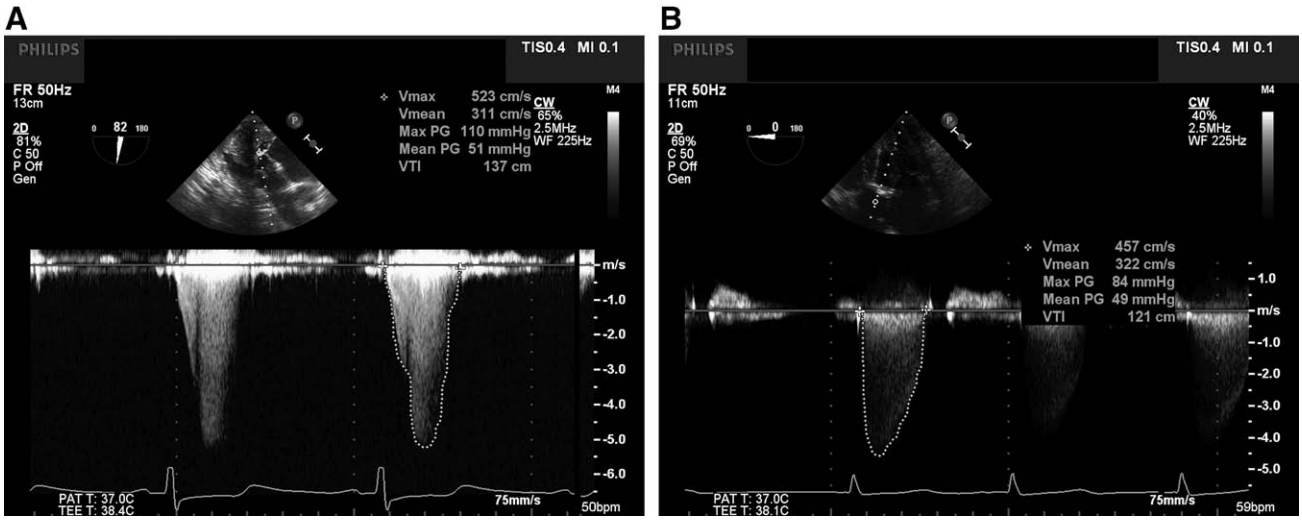

Figure 3. A, Mid- to late-peaking systolic, dagger-shaped profile in a hypertrophic cardiomyopathy patient (continuous wave Doppler; transgastric long-axis (TG-LAX) view, transesophageal echocardiography (TEE) consistent with a subvalvular, dynamic left ventricular outflow obstruction. B, Continuous wave Doppler (CWD) of severe aortic stenosis patient with fixed obstruction. Early systolic peaking parabolic-shaped profile of severe aortic stenosis patient. Deep TG-LAX view. TEE image.

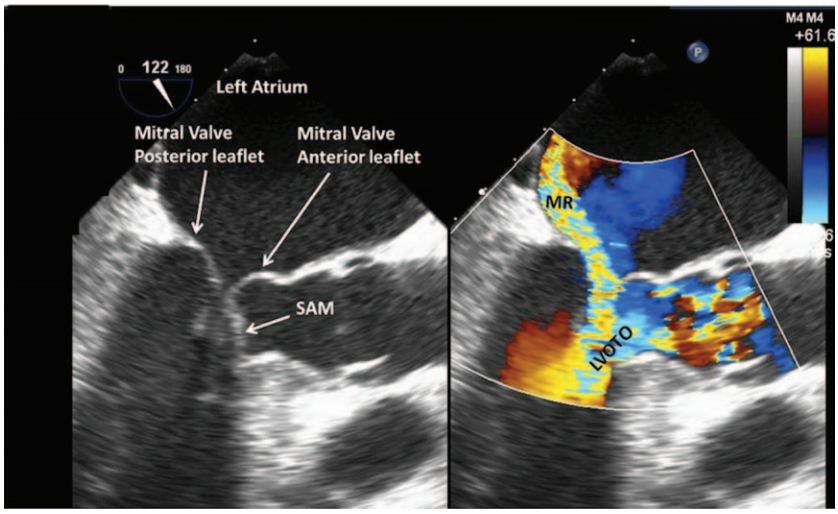

Figure 4. Systolic anterior motion (SAM) of the mitral valve without and with color Doppler. Transesophageal echocardiography (TEE) image. LVOT = left ventricular outflow tract.

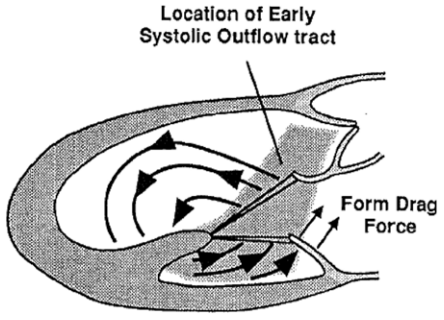

Figure 5. Flow-drag-flow forces on anteriorly displaced posterior papillary muscle cause left ventricular inflow to occur along the septum, while outflow moves from the posterior wall toward the left ventricular outflow tract (LVOT), dragging the mitral valve leaflets anteriorly toward the LVOT. This increases the risk of systolic anterior motion and outflow tract obstruction.

declines because it is approximately proportional to the square of velocity. Next, for any given shape, the ratio of lift to drag decreases with decreased velocity.[21] The numerous studies that showed SAM was occurring at low or normal LVOT velocities and that HCM patients prone to SAM had more anteriorly positioned papillary muscles forcing diastolic inflow and systolic outflow hydrodynamics to change demonstrated drag was the predominant force.[59] In a study of 721 patients, Maron et al.[60] found that 97% of patients with HCM were noted to have SAM.

Importantly, patients with HCM may have intrinsic mitral disease and up to 10% to 20% of patients with HCM have significant MR, which is independent of SAM.[19,39] Jets due to primary leaflet pathology usually occur throughout systole and are often directed anteromedially or centrally.[24] In patients with dynamic LVOTO, the MR jet is classically directed posteriorly and laterally[49] (Fig. 4). Moreover, MR resulting from SAM peaks in mid- to late systole as opposed to throughout systole in those with intrinsic MV disease.[24] SAM is not pathognomonic of HCM and can also develop in up to 6% of patients after mitral repair surgery.

There is an important group of patients who develop dynamic LVOTO and SAM in the absence of HCM in which echocardiography is critical to its diagnosis. These are usually elderly patients with chronic arterial hypertension, LVH, and a sigmoid septum who develop LVOTO in the presence of hyperdynamic LV function due to a number of possible reasons, such as anemia, hypovolemia, systemic vasodilation, and inotropic therapy.[61,62] In a case series of 97 hypertensive patients, factors associated with dynamic LVOTO included female sex, small body surface area and LV cavity size, increased relative wall thickness, LV contractility and resting LVOT velocity, mitral annular calcifications, and SAM of the MV.[62] These patients may present with hypotension, low cardiac output, MR due to SAM, and LA hypertension (e.g., elevated pulmonary artery occlusion pressure per pulmonary artery catheter [PAC]) mimicking LV systolic failure, but in fact have a hyperdynamic, underfilled LV. Therapy is directed at volume expansion, discontinuing inotropic therapy, and often addition of a systemic arterial vasoconstrictor.[62]

TREATMENT AND OUTCOMES—MEDICAL AND SURGICAL

Pharmacologic Management

It has been clearly demonstrated that the presence and magnitude of LVOTO in HCM patients are independent predictors of sudden death and of all-cause cardiac mortality.[53] Considering the mechanisms underlying myocardial contraction (calcium ions binding to troponin C and excitation-contraction coupling) and that obstruction in HCM is dynamic and lethal dysrhythmias are a prominent feature of HCM, a number of medical regimens have been used in these patients with the goal of modulating the natural history.[6] These have included β-blockers, Ca^{++} antagonists (verapamil or diltiazem),[63] and disopyramide regimens, each of which has been demonstrated to be effective compared with no treatment in most HCM patients.[6,21,63–68]

The goal of medical management with these drugs is to lessen or eliminate the LVOT gradient through negative inotropy. Most of these drugs used to treat symptomatic HCM patients to improve their symptoms do so by lessening or eradicating the LVOT pressure gradient. β-adrenergic blockade can improve symptoms of dyspnea and angina due to its decreasing of the dP/dT (change in pressure with respect to time), and thus most likely causing a decrease in LVOTO.[69] Disopyramide has been shown to reduce LVOT pressure gradients, thus increasing survival.[69] It is a weak calcium channel blocker; however, its mechanism of decreasing contractility is through sodium/calcium exchange.[69] Pollick et al.[70] administered disopyramide to 43 HCM patients and found that the LVOT gradient was eliminated or reduced in 100%.

These medications also decrease LVOTO during exercise by blunting the sympathetic response[71] and are useful in treating the symptoms and attenuating the risk of sudden cardiac death. The medications should be titrated based on relief of symptoms and attempting to avoid any significant adverse effects associated with the drugs. More recently, perhexiline, which augments myocyte energy supply, has been shown to improve diastolic dysfunction and symptomatology.[72]

Alcohol Septal Ablation

The technique of alcohol septal ablation using selective engagement of septal coronary branches[21,68,72–76] was first described in 1994 to treat HCM with results comparable with those after surgical resection.[73] Ethanol infused into the septal branches of the left anterior descending coronary artery induces a targeted septal myocardial infarction (MI).[69] Many experts and centers choose to use this approach in patients with significant comorbidities who may not tolerate surgery, while surgical myectomy is reserved for patients undergoing concurrent cardiac procedures and/or in those patients without significant comorbid issues. Additional criteria for alcohol septal ablation are that patients have the following: (1) symptoms that interfere with lifestyle despite medication optimization; (2) septal thickening ≥15 to 16 mm; (3) peak LVOT gradients ≥30 mm Hg at rest or ≥50 mm Hg provoked; (4) accessible septal branches; and (5) absence of abnormalities of the MV and of proximal left anterior descending coronary artery stenosis or severe coronary artery disease. These are essentially the same criteria for surgical septal myectomy without the last 2 criteria.[69]

Alcohol septal ablation is not without potential problems. The most common complication is a right bundle branch block, which has a postprocedural incidence of approximately 50%.[77] Other complications include coronary dissections, remote MI due to collateral circulation or an ethanol injection into the incorrect coronary, ventricular septal rupture, heart failure, and heart block.[69] In a recent nonrandomized study of 177 patients undergoing alcohol septal ablation with a 5.7-year follow-up, survival free of all mortality was comparable with age- and sex-matched controls of surgical myectomy patients (79% vs 79%; $P = 0.64$).[73] Even though selection criteria for surgical myectomy and alcohol septal ablation are similar, the "gold standard" for first consideration is still surgical septal myectomy, when performed in experienced centers in patients who are drug-refractory and severely symptomatic.[6,78] The echocardiography guidelines for alcohol septal ablation are outside the scope of this review; they can be found in the American Society of Echocardiography Clinical Recommendations for Multimodality Cardiovascular Imaging of Patients with Hypertrophic Cardiomyopathy.[31]

Table 5. History and Evolution of Surgical Myectomy for Hypertrophic Cardiomyopathy

1958	Cleland performs first myectomy through aortic root in Great Britain[9]
1959	Kirklin and Ellis perform first myectomy through ventriculotomy approach at Mayo Clinic[7]
1960	Morrow and Brockenbrough first publish report of 2 cases performed through ventriculotomy that subsequently has borne Morrow's name[8]
1990s	Messmer performs extended Morrow myectomy, which included more midventricular muscle
2000s	Swistel developed resection-plication-release procedure,[79] which involves the plication and reduction of large anterior leaflet and release by dividing any abnormal attachment of papillary muscle to lateral wall of ventricle

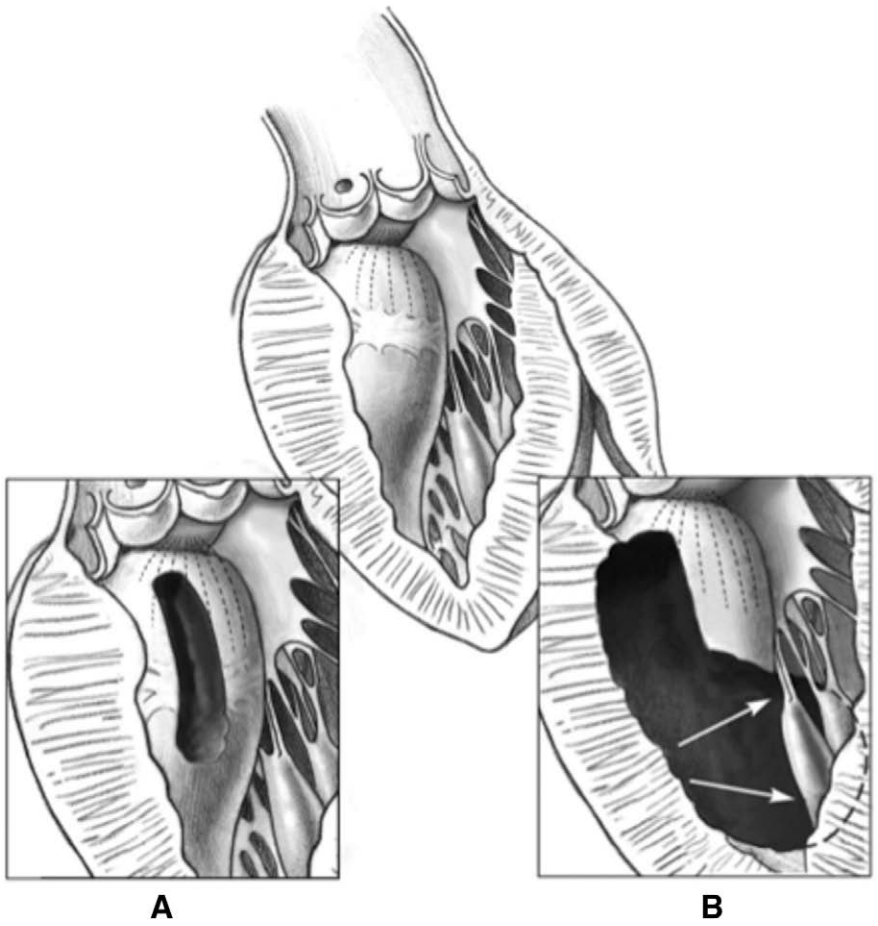

Figure 6. Comparison of the classic Morrow procedure (A) versus the extended septal myectomy modification (B). Resection of the septal wall is extended toward the apex, toward the free wall on the left side of the image, and then toward the right, as indicated by the white arrows. The dashed lines in the basal septum represent fascicles of the left bundle originating from the membranous septum.[80]

Surgical Management

Since the time of the first myectomy through the aortic root by Cleland in Great Britain in November 1958, the surgical procedure has evolved (Table 5).[8,79,80] Figure 6, A and B demonstrates the classic Morrow myectomy versus the extended Morrow myectomy.[80] This technique progressed with Swistel and his resection (extended myectomy), plication (horizontal mitral plication), and release (manipulation of subvalvular structures) (Fig. 7).[79] The American and European Colleges of Cardiology recommend myectomy in (1) patients with labile obstruction and peak LVOT pressure gradients ≥50 mm Hg during exercise or provocation and resting gradients >30 mm Hg and (2) patients with NYHA class II through IV symptoms refractory to medical therapy.[10]

TEE is essential to guide surgical myectomy, assess the myectomy and/or MV repair, and exclude potential complications. TEE provides important information about residual gradients, obstructive SAM, septal anatomy, and the need for immediate postmyectomy revision. It is essential to attempt provocation of LVOTO to ensure adequate muscle resection. The 2 most common methods are (1) response to induced premature ventricular contraction (PVC) to assess postextrasystolic potentiation and (2) administration of isoproterenol.[81] PVCs, which are weaker than a regular contraction, lead to provocation by causing an increase in contractile strength in several regular beats that follow it.[82] Isoproterenol is used for enhancement due to its β-1 and β-2 agonist properties. It is a superior inotrope to use over others because it causes increased contractility, decreased afterload and preload through arterial and venous dilation, and tachycardia. All these physiologic responses should induce LVOTO by simulating exercise.[83] An adequate response to isoproterenol (starting at 1 mcg/kg/min) would result in an increase in the heart rate to >120.[81] Patients with resting gradients >25 mm Hg or post-PVC provoked gradients >50 mm Hg or greater than moderate MR should have prompt revision.[84–86] In some cases, gradients measured by TEE may be inaccurate, and intraoperative direct measurement of gradients by the surgeons can give more accurate gradient information.[87]

Ventricular septal defects (VSDs) are 1 potential complication of surgical myectomy, and TEE is vital in determining if this has occurred. Septal thickness <20 mm before myectomy increases the risk of VSD, as well as elderly patients and those who are simultaneously having coronary artery bypass grafting.[84] Other complications include severed septal perforator arteries, aortic insufficiency, and heart block. TEE findings that include a color jet in the LVOT after myectomy should arouse suspicions of aortic insufficiency, severed septal perforator, or VSD.[88] The mechanism of immediate postmyectomy aortic insufficiency is likely secondary to excision of the supporting septal wall musculature that gives structure to the aortic valve.[89]

Overall, periprocedural mortality has improved dramatically since the introduction of septal myectomy and in highly experienced centers is 1% to 2%.[90–93] Increasing age

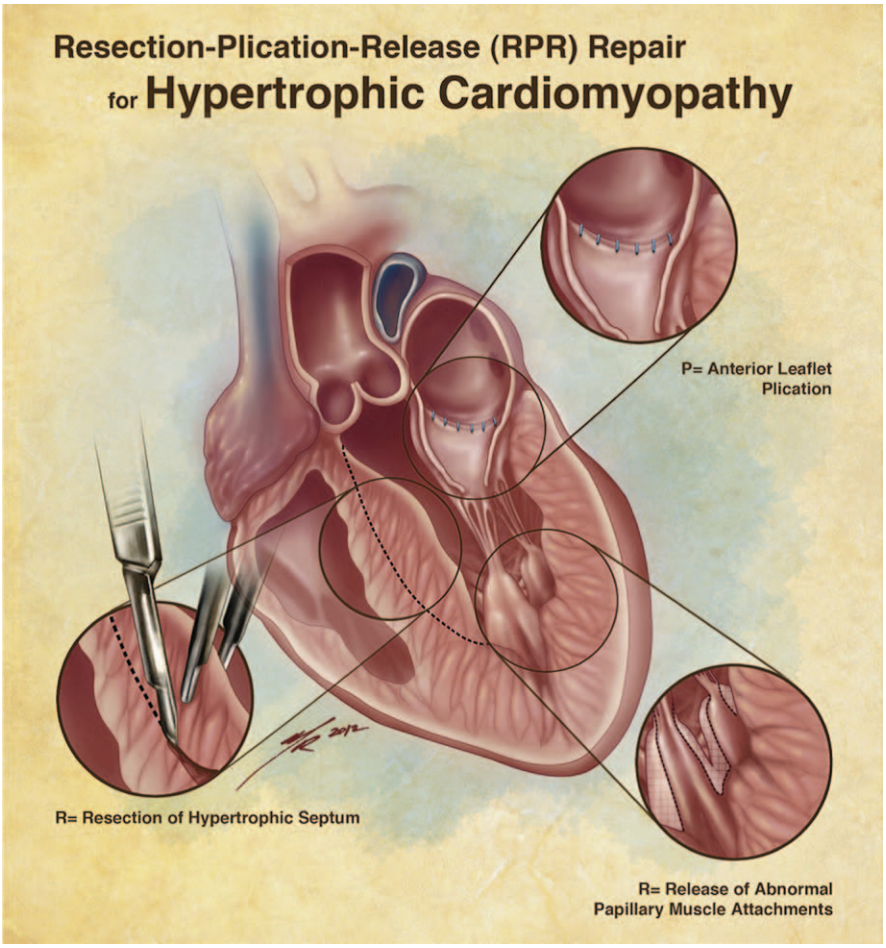

Figure 7. Schematic representation of the hypertrophied heart in hypertrophic cardiomyopathy, depicting the morphologic variant leading to obstruction and potential surgical options for management including resection (extended myectomy), plication (horizontal mitral plication), and release (manipulation of subvalvular structure).[79]

and concurrent coronary artery disease portends a worse long-term outcome.[94]

In addition to myectomy, HCM patients may be candidates for implantable cardioverter-defibrillator (ICD) implantation. The following are indications for ICD placement: (1) positive family history of premature sudden cardiac death as a result of HCM[74]; (2) documented nonsustained ventricular tachycardia[75]; (3) syncope at rest or during exercise; (4) LVH >30 mm[35]; and (5) an abnormal arterial blood pressure response to exercise, which can be described as an increase in systolic blood pressure of <20 mm Hg from the baseline value or a progressive decrease in the systolic value by 20 mm Hg after an initial increase.[76,95]

Mechanisms Underlying Clinical Improvement

Although still considered controversial, the postsurgical decrease in LVH seems to be a critical determinant of prognosis.[35,96,97] When evaluating patients after septal myectomy, Monteiro et al.[98] demonstrated significant reductions in septal and posterior wall thickness, as well as improved diastolic filling. In a retrospective analysis of postoperative patients, Deb et al.[99] describe a significant regression of LVH as early as patient discharge, especially in patients younger than 50 years. This regression was sustained over 2 years and was estimated to be 15 times the mass of muscle removed at surgery.[99]

A multicenter retrospective study of 1337 patients followed 3 patient subgroups: patients with LVOTO who had undergone myectomy at the Mayo Clinic, a specialized HCM center, those with LVOTO who had not undergone myectomy, and HCM patients without LVOTO. The nonoperative HCM patients in this study were compiled from 3 other centers that did not have specialized surgical experience in HCM. In HCM patients who had had a myectomy, NYHA functional class was significantly improved (2.9 pre vs 1.5 post, $P < 0.001$). Moreover, the operative mortality was 0.8% and the 1-, 5-, and 10-year survival rates were 98%, 96%, and 83%, respectively (Fig. 8).[100] These long-term survival rates were significantly better than in the other 2 groups.[100]

Single-center studies have also demonstrated improvements in mortality/symptomatology after myectomy. Schulte et al.[101] demonstrated improvements in NYHA functional class. Other studies have demonstrated improvements in exercise performance after myectomy.[102]

HCM PATIENTS UNDERGOING NONCARDIAC SURGERY

Preoperative Management

Unless there are signs of a pathologic murmur (systolic murmur radiating to the neck) or unexplained cardiac signs (rales, significant peripheral edema) or symptoms (exertional dyspnea, angina, or syncope), patients who have underlying HCM may go undiagnosed during the preoperative evaluation, especially if there are no known family members with HCM. In the absence of symptoms and

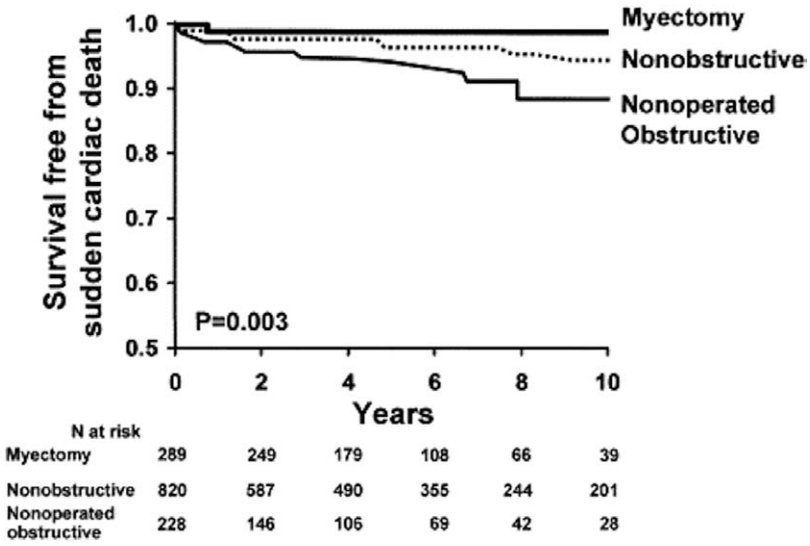

Figure 8. Survival free from hypertrophic cardiomyopathy-related death among patients in 3 hypertrophic cardiomyopathy (HCM) subgroups: surgical myectomy (n = 289), nonoperated with obstruction (n = 223), and nonobstructive (n = 820). Overall log-rank, $P < 0.001$, myectomy versus nonoperated obstructive HCM, $P < 0.001$; myectomy versus nonobstructive HCM, $P = 0.01$.[100]

Table 6. Clinical Features of Functional (Benign) Heart Murmurs

-Location: left sternal border and nonradiating[a]
-Timing: mid- or early systolic[b]
-Intensity: grade 2 or lower
-No unexplained cardiac or pulmonary symptoms (e.g., dyspnea, chest pain, orthopnea, syncope)
-No additional unexplained cardiac signs (e.g., rales, S3, significant peripheral edema)
-No electrocardiographic or chest radiographic evidence of ventricular hypertrophy)

HCM = hypertrophic cardiomyopathy.[26,103]

[a]Murmurs radiating into the neck should be considered due to aortic stenosis or HCM and are thus not functional.

[b]Diastolic murmurs are always considered pathologic.

signs, ECG findings (Table 1) may suggest that a patient has underlying HCM.

During physical examination, murmurs should be evaluated for dynamic changes, and patients with murmurs that do not meet the criteria of a benign murmur (Table 6)[103] should be referred for echocardiographic examination before surgery. Dynamic changes in the murmur intensity can be assessed by determining the modulating effects of the Valsalva maneuver, standing and exercise that increase bruit intensity or, conversely, squatting and β-blockade that decrease intensity. It is important to know a patient's baseline ECG (Table 1) to monitor intraoperative changes and their correlation with hemodynamic perturbations. However, these common ECG abnormalities do not correlate with severity or pattern of hypertrophy.

Patients with diagnosed HCM can present with any number of symptoms, ranging from minimal to those consistent with end-stage heart failure. Mild symptoms are defined as dyspnea or palpitations on significant exertion, whereas severe symptoms include syncope with exertion, orthopnea, palpitations that cause presyncope or syncope, and peripheral edema in the lower extremities.[104] Symptomatology of the patient will guide the choice of intraoperative monitoring; therefore, it is essential to determine the patient's NYHA stage of congestive heart failure if he or she is experiencing symptoms. Family history is valuable, but even more significant is the patient's own symptoms and functional class. Initial patient evaluation should attempt to determine disease severity, by assessing functional status, cardiac and respiratory symptomatology, personal and family cardiac history, current medications, history of rhythm disturbances and previous strokes, or congestive heart failure. Patients should be instructed to continue β-blockers, calcium channel blockers, and disopyramide and maintain proper hydration perioperatively.[16,105] Although rare, some HCM patients with a history of excessive bleeding may actually have an acquired von Willebrand disease due to dynamic LVOTO-related shearing of large multimers of von Willebrand factor.[106] Mechanistically, this is identical to the Heyde syndrome described in the setting of valvular AS.[107] This bleeding propensity may be important depending on the type of surgery and anticipated blood loss.

Preoperative echocardiographic assessment should include determination of abnormalities of the MV and subvalvular apparatus, MR, LVOT gradients, LV systolic function, degree of diastolic dysfunction, chamber enlargement, and results of prior interventions (percutaneous septal ablation or septal myectomy). Moreover, the presence of an automatic ICD and if it has been recently interrogated should be determined. Atrioventricular node integrity and underlying rhythm evaluation are critical in patients who have had either septal myectomy or ablation and in patients with a pacemaker in situ. Most of these data can be determined via an echocardiographic evaluation. The critical information gleaned from the echocardiographic evaluation include (1) the LVOT pressure gradient (significant if >30 mm Hg peak gradient at rest or >50 mm Hg peak provoked gradient), (2) LV systolic function, and (3) LV diastolic function (Table 7).

MV abnormalities are observed in two-thirds of patients with HCM, but antibiotic prophylaxis is not required regardless of MR.[108] Decisions regarding postoperative disposition, including intensive care justification, should consider disease severity and the specifics of the planned surgical procedure. Adverse cardiovascular outcomes, including myocardial ischemia, arrhythmias, and congestive heart failure, are

Table 7. Echocardiographic Red Flags for Hypertrophic Cardiomyopathy Patients Undergoing Noncardiac Surgery due to Increased Risk for Perioperative Complications

- Significant LVOT pressure gradients (>30 mm Hg peak gradient at rest or >50 mm Hg peak gradient under provocation)
- Moderate-severe MR due to systolic anterior motion of the mitral valve (SAM) (vena contracta 0.3–0.69 mm = moderate MR; ≥ 0.7 mm = severe MR)
- LV systolic function depressed with EF <45%–50%
- Restrictive diastolic dysfunction by TMDF and pulmonary venous Doppler flow (PVDF) velocity profile [early-to-late TMDF velocities (E/A >1.7); isovolumic relaxation time (IVRT) <90 ms; deceleration time (DT) <140 ms; PVDF systolic velocity to diastolic velocity (S/D) <0.8

LVOT = left ventricular outflow tract; TMDF = transmitral Doppler flow.

determined primarily by the severity of the underlying HCM/LVOTO and the perioperative management.[109]

Intraoperative Management

Up to 60% of patients with HCM experience perioperative cardiac events, for example, MI, congestive heart failure, or both.[110] In a retrospective chart review study, HCM patients undergoing noncardiac surgery had increased rates of both MI and death. HCM patients had an overall absolute mortality rate 4.2% higher than the control group[23]; therefore, understanding the physiologic goals for these patients is imperative. There are no published guidelines for anesthetic management of HCM patients, and a literature review yields only cohort studies and case reports of complicated patients.[23,103–105,109–114]

Sympathetic Stimulation and Increases in Contractility

The hemodynamic goals in managing patients with HCM are similar to those of an AS patient with regard to maintaining afterload, a slower heart rate, and sinus rhythm. The most significant difference is the dynamic nature of HCM and associated LVOTO. This is in contrast to fixed lesions like AS. Increases in heart rate, rhythm disturbances, and decreases in afterload will exacerbate LVOTO and may cause hemodynamic deterioration. In addition, increases in contractility and decreases in preload will accentuate LVOTO, highlighting the dynamic component of HCM. Therefore, the principles of treatment for hypotension are volume expansion (including increasing preload in the Trendelenburg position) and drugs that increase systemic vascular resistance (SVR) without increasing heart rate or contractility (e.g., phenylephrine and vasopressin). Sympathetic stimulation secondary to patient anxiety, intubation, and surgical incision and acute changes in preload, afterload, and contractility secondary to the pharmacological effects of anesthetic drugs, surgical blood loss, and postoperative pain can precipitate hemodynamic collapse. It is also imperative to maintain sinus rhythm because LVH and decreased compliance in HCM dictate increased dependence on atrial kick for maintaining the cardiac output. Direct current cardioversion may be necessary in the case of sudden onset of atrial fibrillation that is hemodynamically unstable.[16]

Although both general and neuraxial anesthesia can be used, it is important to have a clear understanding of the hemodynamic changes associated with each option. Depending on the IV or inhaled drugs chosen, close monitoring and management of the medication's modulating effect on afterload, preload, contractility, heart rate, and sympathetic activity are critical. Neuraxial techniques may also be considered. In general, a slow controlled titration of medication via an epidural is preferred over a spinal with the goal of maintaining afterload and preload and avoiding sympathetic stimulation.

Several monitoring tools should be considered when managing HCM patients. In choosing monitoring modalities, the anesthesiologist should take into consideration the following: (1) degree of LVOTO (obstruction at rest, labile obstruction, or no obstruction); (2) whether or not the patient has signs or symptoms of heart failure; (3) the type of surgery and potential for large volume loss or fluid shifts; and (4) other comorbidities that place them at higher risk of perioperative complications (i.e., diabetes, renal insufficiency). In addition to the standard American Society of Anesthesiologists monitors, an intraarterial catheter and/or noninvasive pulse plethysmographic variability (PPV) index monitor and central venous pressure (CVP) monitoring may be considered.[115] In 2 meta-analyses, PPV index monitors were shown to be a reasonably reliable indicator of preload responsiveness in perioperative and critically ill patients.[116,117] In patients spontaneously breathing or in those who have arrhythmias, PPV index monitors are not as trustworthy. The value and reliability of this preload-responsiveness monitor has not been evaluated in HCM patients. PPV index monitors may be beneficial in the low-risk HCM patient (nonobstructor) undergoing low-risk to intermediate-risk surgery. Intraarterial catheters should be considered in those HCM patients with labile obstruction or resting obstruction, especially if undergoing intermediate- to high-risk noncardiac surgery. Sudden hypotension should be recognized and treated promptly by maneuvers that attenuate and prevent LVOTO. Central venous lines may be useful in HCM patients by providing oxygen delivery and demand information through central venous oxygen saturation, rapid central access for administration of vasoactive drugs, and evaluation of dynamic changes in the CVP particularly as it relates to volume changes and responsiveness. However, according to 2 meta-analyses in both critical care and intraoperative patients (not HCM patients), CVP is not a reliable indicator of blood volume nor is the CVP/delta CVP predictive of hemodynamic responsiveness to fluid challenges.[118,119] Thus, the risks of placing such catheters may outweigh the benefits but should be evaluated on a case-by-case basis. For an HCM patient (especially those with labile obstruction) undergoing intermediate- or high-risk noncardiac surgery that will involve the potential for large volume losses, a PAC may be considered. It may be helpful to establish a baseline pulmonary artery wedge pressure (filling pressure) and cardiac outputs by thermodilution to follow a trend to assist in managing an HCM patient having LVOTO. In the event of hypovolemia or decreased SVR causing acute LVOTO, the pulmonary artery wedge pressure tracing may demonstrate a decrease in the cardiac index or even the sudden appearance of large V waves (indicating LVOTO, SAM, and MR). However, large V waves are neither sensitive nor specific for acute MR.[120]

Last, the overall hemodynamic goals include maintaining the mean arterial blood pressure at >65 to 70 mm Hg to maintain coronary perfusion pressure to the subendocardium in the hypertrophied heart.

Likely, the most useful monitor for high-risk patients undergoing high-risk surgery is TEE. TEE can determine whether hemodynamic perturbations are caused by hypovolemia, increased LVOTO or SAM, or LV systolic dysfunction. TDI can assist in determining LV filling pressures in patients whose lungs are mechanically ventilated, with E′ representing myocardial elongation during early diastole and relating to the velocity of relaxation.[121–123] Combes et al.[121] found that E/E′ >7.5 predicted a pulmonary artery occlusion pressure of 15 mm Hg or higher with a sensitivity of 82% and specificity of 81%. The predictive accuracy of transmitral Doppler flow velocities in HCM patients with abnormal myocardial relaxation has been reported. Geske et al.[124] investigated 100 symptomatic patients with HCM who underwent measurement of E/E′ simultaneously (n = 42) or within 48 hours of cardiac catheterization and direct LA pressure measurements. There was statistically significant correlation between Doppler-derived diastolic velocities and invasive measurements. However, the predictive accuracy of E/E′ for estimation of LA pressure in the individual patient was moderate.[124]

HCM AND THE PARTURIENT

Krul et al.[125] have provided a systematic review of pregnancy in women with inherited cardiomyopathies including HCM. HCM is not a contraindication to pregnancy, and asymptomatic patients or those with mild symptoms typically tolerate pregnancy well. Physiologic changes in pregnancy that have a salubrious effect on the pathophysiology of HCM include an increase in total blood volume (50% increase in plasma volume and 30% increase in red blood cell mass) and increased LV end-diastolic diameter associated with increased stroke volume.[126] The increase in blood volume offsets the other physiologic changes in pregnancy, which could aggravate HCM such as increased cardiac contractility and decreased SVR, in part due to the low-resistance placenta. Autore et al.[127] have found that cardiac morbidity during pregnancy in women with HCM is closely correlated with NYHA functional status before pregnancy, and the incidence of complications during pregnancy depends on the patient's baseline functional status. In a retrospective review of 127 HCM parturients and assessing 271 pregnancies over 40 years, pre-pregnancy symptoms worsened in only 10%, with dyspnea being the most frequent.[111] In this cohort, there were 2 primigravidas who developed pulmonary edema 5 days postpartum. One patient had general anesthesia for obstetric reasons, and a review of the record did not show evidence of excessive IV fluids. The second patient had an epidural for pain management and a normal vaginal delivery. This patient received IV fluid after hypotension.[111] Patients at high risk for sudden cardiac death are those with previous episodes of sustained ventricular tachycardia or those who have a strong family history of sudden cardiac death.[127] This is not different compared with nonpregnant HCM patients. Even though only small retrospective studies have demonstrated HCM parturients with mild symptoms tolerate pregnancy well, it is strongly recommended that these patients be transferred to a tertiary care high-risk obstetrics center.

Hemodynamic instability is most likely to occur during labor due to increased sympathetic activation from pain and anxiety causing tachycardia, recurrent decreases in venous return from bearing down (Valsalva maneuver), uterine contractions, and fluid shifts after the third stage of labor. Immediately postpartum, the cardiac output increases by up to 75% above predelivery values and then slowly declines over the next 2 weeks. The increase in stroke volume may decrease the risk of LVOTO and offset the blood volume lost at delivery (approximately 1000 mL for cesarean delivery and 500 mL for vaginal delivery). During this peripartum period, patients with established diastolic dysfunction and HCM are at risk of LVOTO, arrhythmia, or pulmonary edema.[125]

Although neuraxial anesthesia will decrease afterload, there are reports of such techniques being used successfully in this subset of patients.[125] Pryn et al.[128] report the successful use of a combined spinal and epidural technique in HCM parturients undergoing cesarean deliveries. They reported using 1 mL of 0.5% hyperbaric bupivacaine for the spinal after 1 L of crystalloid was administered, followed by incremental dosing of 0.5% bupivacaine through the epidural to obtain a T3 sensory block. Two percent lidocaine injected slowly and incrementally through an epidural catheter to obtain T4 sensory block after volume loading has also been reported.[112] Okutomi et al.[129] describe using a slow continuous spinal infusion of 25 mcg fentanyl and 50 mcg epinephrine/hour diluted in dextrose 5% for a vaginal delivery. Hemodynamic management after a spinal anesthetic may be more challenging, and slow titration using a continuous spinal, decreased dosing of intrathecal medication, advanced fluid loading, and patient positioning are critical aspects to this management.

Cesarean delivery should only be performed for obstetric indications, decompensated heart failure, or symptomatic LVOTO,[125] and an intraarterial catheter for monitoring is recommended. CVP monitoring and/or PACs may be considered, especially in those with decompensated heart failure. These can be used to assess filling pressures and cardiac outputs, as well as mixed venous/central venous oxygen saturations, and provide for rapid delivery of vasoactive medications. Again, the risk/benefit ratio should be assessed and level of experience in placement of PACs and interpreting the data. TTE or TEE capability may be helpful in the event of hemodynamic instability.[130]

Table 8 provides a summary of the recommended management before and during pregnancy for parturients with HCM.

FUTURE RESEARCH PRIORITIES

Currently, treatments for HCM are directed at symptomatic relief and preventing sudden death. The future goal of research is focused on changing the natural course of the disease and preventing its phenotypic expression. Specific initiatives have been identified by the working group of the National Heart, Lung, and Blood Institute to assist in the development of novel treatments and include clinical, translational, and basic science aspects of the disease.[131] Clinical initiatives involve defining all the genetic causes of HCM and the natural history of the disease, as well as encouraging more randomized controlled trials involving

Table 8. Recommendations for the Management of Women with Hypertrophic Cardiomyopathy Before and During Pregnancy

Before pregnancy
Assessment of symptomatology
Echocardiography (LV systolic and diastolic function, LVOTO, mitral regurgitation)
Exercise testing
Start medication (β-blocker, preferably metoprolol) when symptomatic
Risk assessment and counseling
Genetic counseling
During pregnancy
Follow up each trimester (low-risk) or monthly/bimonthly (increased risk)
Clinical assessment, echocardiography
Continue β-blocker or start when symptomatic
Prompt treatment of atrial fibrillation
Delivery
Vaginal delivery appropriate
Blood pressure and ECG monitoring
Consider placement of central venous or pulmonary artery pressure monitors, especially if patient shows signs of decompensated CHF
Monitor preload echocardiographically when necessary
Cautious use of epidural or spinal anesthesia
Replace fluid loss, avoiding overhydration
Oxytocin only as slow IV infusion due to dramatic decrease in SVR if given as a rapid bolus
Clinical observation for at least 24 h post delivery

LVOTO = left ventricular outflow tract obstruction; ECG = electrocardiography; CHF = congestive heart failure; SVR = systemic vascular resistance.[130]

treatments. Translational research initiatives focus on understanding the mechanism of the disease and involve attempting to prevent expression of mutant alleles, describing the progression of pathologic HCM phenotypes, and defining the impact of arrhythmias on the disease and patients with HCM. Finally, basic science research is directed toward identifying the stimuli activated by the expression of mutated sarcomeres, looking for matrix and nonmyocyte cells contributing to the mutant expression and the role of vasculature in the disease process, as well as searching for altered myocyte metabolism in the presence of HCM. Given that nearly 1 in 500 adults have HCM and it is a leading cause of sudden death in young people, there is a clear need for better treatments and understanding of the disease, and this initiative will hopefully provide organization and progress toward this goal.[131]

CONCLUSIONS

The diagnosis of HCM is based predominantly on echocardiographic variables including dynamic LVOTO, LVH, distribution of increased muscle thickness, mechanism and severity of MR, and degree of diastolic dysfunction. Current indications for surgical intervention include symptomatic patients refractory to medical therapy who can tolerate the risk of surgery and patients with marked outflow gradients, even if asymptomatic. Although unclear, the mechanism underlying improvement in symptoms, LVOTO, and long-term survival after myectomy is at least in part due to LVH regression. It is crucial for anesthesiologists to understand the pathophysiology of this disease to best manage these patients in the perioperative setting.

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